Role of noninvasive ventilation in perioperative patients with neuromuscular disease: a clinical case

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Received 5 March 2013; accepted 10 June 2013
Available online 6 April 2014

Abstract The inclusion body myositis is an inflammatory myopathy that leads to chronic muscle inflammation associated with muscle weakness. It is characterized by a restrictive ventilatory syndrome requiring ventilatory support under non-invasive ventilation. The authors describe a clinical case and the anaesthetic management of a patient with inclusion body myopathy candidate for vertebroplasty, which highlights the importance of locoregional anaesthesia and of noninvasive ventilation and includes assisted cough techniques, maintained throughout the perioperative period.

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PALAVRAS-CHAVE
Ventilação não invasiva; Anestesia locorregional; Miosite por corpos de inclusão

Papel da ventilação não invasiva no período perioperatorário de doentes com patologia neuromuscular: caso clínico

Resumo A miosite por corpos de inclusão é uma miopatia inflamatória que cursa com inflamação crônica muscular associada à fraqueza muscular. Caracteriza-se por uma síndrome ventilatória restritiva com necessidade de suporte ventilatório sob ventilação não invasiva. Os autores descrevem caso clínico e respectivo manuseio anestésico de paciente com miopatia por corpos de inclusão proposta para vertebroplastia que realça a importância da anestesia locorregional e da ventilação não invasiva e inclui as técnicas de tosse assistida, mantidas durante todo o período perioperatorário.

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cough and accumulation of oropharyngeal secretions imply high risk of failure in tracheal extubation after positive pressure ventilation in patients with NMD. Therefore, these patients are considered at high risk for general anaesthesia.\(^1\)\(^2\) They are also usually patients with associated comorbidities, which makes them preferred candidates for regional anaesthesia techniques.\(^3\)

The inclusion body myositis is a rare disease that is part of a group of muscle diseases known as inflammatory myopathies, which are characterized by chronic muscle inflammation associated with muscle weakness. Decreased muscle strength is usually progressive (occurs gradually over months or years) and affects both proximal and distal muscles, and can affect only half the body. It is more frequent in males and its symptoms usually begin after age 50. The familial forms usually occur in childhood and show no inflammatory changes. Both forms have intracytoplasmic and intranuclear inclusions in muscle tissue.\(^4\) There is neither a cure for this disease, nor a clear routine of care.

The involvement of the respiratory muscles, characterized by a restrictive ventilatory syndrome of progressive evolution, leads to the need for ventilatory support under non-invasive ventilation (NIV). Given the acknowledged efficacy of NIV associated with the techniques of assisted cough in NMD, these should be continued in the perioperative period in patients who were previously under NIV and thus simplify both surgery and the anaesthetic act.\(^5\) The mechanical ventilation ensures better stability of the breathing physiology, with fulfilment of gas exchange, thereby reducing the effects of surgical stress.\(^4\)

**Clinical case**

Female patient, 71 years old, accompanied in lung consultation due to inclusion body myositis, with anaesthetic risk ASA III (concomitant comorbidities: hypertension, atrial fibrillation); ventilation about five years ago with BiPAP at home (18 h/day), with indication for percutaneous kyphoplasty in reason of osteoporotic (or post-traumatic) fracture of T11–T12.

Considering that ours was a patient at high anaesthetic risk, given her lung function (forced vital capacity = 1.05 L, with loss of 22% with dorsal decubitus and peak cough flow of 120 L/min cough – severe restrictive syndrome with frank reduction of peak cough), we opted initially for a medical treatment with analgesia and immobilization in hospital for 12 days.

Despite this treatment, the patient persevered with intense pain complaints; thus, after a multidisciplinary discussion of her clinical case, we decided to proceed with percutaneous kyphoplasty under intraoperative NIV, with the support of mechanical in-exsufflator to ensure the removal of secretions that could eventually arise during surgery.

To improve the conditions in the preoperative, assisted cough was initiated and taught to the nursing staff and to patient’s relatives in the week before surgery, with improvement in ventilatory parameters according to nocturnal oximetry and arterial blood gas.

Then, the surgery was performed under thoracic epidural anaesthesia, at the level T12-L1, with administration of 60 mg of 0.75% ropivacaine. A higher level of superior sensory block in T7 was achieved, and the surgery proceeded with the patient positioned in ventral decubitus. In the operating theatre not only were present the medical teams of orthopaedics and anaesthesiology, but also of pulmonology, to safeguard the eventual possibility of a required intubation by fibreoptic bronchoscopy (in reason of her positioning) and consequent invasive ventilation.

Throughout the surgery the patient remained under NIV by bi-level positive airway pressure (BiPAP) with a nasal mask, at pressures of 5 cm H\(_2\)O for expiratory positive airway pressure (EPAP) and 18 cm H\(_2\)O for inspiratory positive airway pressure (IPAP) in spontaneous/timed (ST) module, with no need for additional assisted cough techniques intraoperatively. The patient always remained without pain, hemodynamically stable, and with no episodes of desaturation, shortness of breath or accumulation of secretions along about 120 min of duration of the procedure. It is noteworthy that there was no need for supplemental oxygen. The peripheral oxygen saturation always remained above 97% under room air ventilation. In the Post-Anaesthetic Care Unit, due to desaturation and the patient complaint of a feeling of presence of bronchial secretions, we used the assisted cough. The episode reversed immediately after removing the plug of mucus secretions. In the absence of additional complications, the patient was discharged to the patient orthopaedics ward after a few hours of recovery, and during this time she remained uneventful until the date of her hospital discharge.

Preventive measures on the ambulation of the patient at home were taken. The use of a wheelchair was encouraged and the patient maintained clinical criteria for the continuous use of assisted cough techniques at home. About two weeks after the surgery, the patient was reassessed in a lung consultation.

**Discussion**

The percutaneous kyphoplasty/vertebroplasty, a minimally invasive technique used in the treatment of osteoporotic fractures and osteolytic tumours of vertebral bodies, consists of a percutaneous introduction of a balloon into the affected vertebra; its inflation and subsequent deflation/removal creates a vertebral space that is subsequently filled with acrylic or biological cement.\(^3\) This is a surgical procedure that involves the placement of the patient in a prone position, and usually the procedure is performed under general anaesthesia.\(^7\) Over time, with the increasing experience of orthopaedists with this technique, it has been possible in many cases to reduce the time necessary for performing this type of surgery. It also has been possible to adjust the anaesthetic technique associated. Thus, if initially nearly all patients were operated under general anaesthesia, recently the sedation has emerged as an increasingly used option, although invariably this technique needs to be supplemented by a good local anaesthesia, particularly for the peristeum.\(^7\) Nonetheless, there are specific times when patients end up feeling some discomfort, especially with introduction of trocars, balloon inflation and injection of cement.\(^3\)

In the present case, taking into account the patient’s comorbidities and the fact that this type of intervention is
still uncommon in the local hospital (and therefore highly likely to take more time), general anaesthesia and sedation were discarded as options. It was then proposed a neuraxial blockade that, in this context, is particularly interesting: this technique avoids the manipulation of the airway and the use of invasive ventilation. Therefore, offers clear advantages, both in intra- and postoperative phases, for patients with NMD and with involvement of underlying respiratory muscles. Recently, Souvatzis et al. reported a case series of patients undergoing kyphoplasty under subarachnoid block (SAB). By being limited in time, SAB is ineffective in prolonged surgical procedures. But the epidural block, thanks to the presence of a catheter, allows, on the one hand, to overcome this setback of a longer surgery than anticipated and, on the other hand, the intraoperative need for kyphoplasty on adjacent vertebrae and allows an adjustment for the adequate anaesthetic level. Additionally, although not described in their paper, the dose of local anaesthetic used by Souvatzis et al. usually limits the occurrence of more significant haemodynamic alterations, compared to the case of epidural anaesthesia – and this is an instability to be avoided in patients at high anaesthetic risk.

The proposition of this anaesthetic procedure, along with ventilation secured by NIV, enabled our patient with NMD to choose a therapeutic option that allowed the immediate resolution of her clinical condition. The patient was discharged from hospital on the third postoperative day, walking with support and without pain. Otherwise, the options would consist of conservative treatment (immobilization and all its associated risks), or kyphoplasty under general anaesthesia under invasive ventilation, which would involve intensive care in the postoperative period and eventual need for tracheostomy and decannulation, entailing a longer period of hospitalization and consequent higher risk of nosocomial disease.

The multidisciplinary evaluation and monitoring throughout the perioperative period, including during the surgical procedure, were critical to the therapeutic success, because situations of eventual need for invasive ventilation or of haemodynamic instability that could arise were guaranteed and safeguarded.

One should also emphasize the importance of the preoperative improvement of the underlying respiratory dysfunction in this type of patient. Throughout the perioperative period, the assisted-cough techniques contribute to minimize the risk of respiratory complications to which these patients are subject to. In the case presented, there was no need for the use of assisted cough techniques during surgery, which was attributed to the extensive use of this technique in the preoperative period.

With the reassessment of the patient on the second postoperative week, it was possible to conclude that her quality of life was superior than that presented before the fracture/surgical treatment, and this clearly demonstrates the benefits of the clinical options chosen.

**Conclusion**

By avoiding the airway manipulation, the epidural anaesthesia offers many advantages for patients with neuromuscular pathology in need of surgery. The multidisciplinary approach and the use of techniques of ventilatory support ensure a better quality of life for these patients; therefore, these should be promoted throughout the perioperative period.

**Conflicts of interest**

The authors declare no conflicts of interest.

**References**